

Spontaneous Resorption of an Occipital Meningocele: Computed Tomography and Magnetic Resonance Imaging Evaluation

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Abstract

Keywords

- meningocele
- computed tomography
- magnetic resonance
- occipital meningocele
- encephalocele
- surgery

Cranial meningocele is a very rare variant of encephalocele. Meningocele can be associated with other disorders and may cause complications. Therapy is usually based on surgical treatment. To our knowledge, we describe the first case of spontaneous resorption of an occipital meningocele in a full-term newborn boy. A full-term newborn was noted to have a large not skin covered, semitransparent cystic lump in the occipital bone. He underwent computed tomography and a diagnosis of meningocele was proposed. After a few hours, the cystic lump spontaneously resorbed. After 1 week the patient underwent magnetic resonance. Histology confirmed the diagnosis.

Introduction

Encephalocele refers to the herniation of intracranial tissue through a defect in the cranium¹; such lesions are called meningoceles when they contain only meninges and meningoencephaloceles if brain tissue is included in the herniated tissue. Cranial meningocele is a very rare variant of encephalocele, and it is thought to be the product of postneurulation disorders.² The sac of the malformation is generally covered by skin lined interiorly by arachnoid and contains only cerebrospinal fluid.²

Meningocele is sometimes associated with Dandy–Walker syndrome, amniotic rupture, cerebellar dermoid tumor or other disorders.^{3–5} Moreover, this disease may complicate with leptomeningitis since the gap due to the herniation could lead to the infiltration by pathological organisms.⁶ The

therapy is based on surgical treatment. Although it has been reported by Nejat and Kazmi⁷ a case of spontaneous resorption of a sacral meningocele occurred at 6 months of age and a case of spontaneous resolution of a traumatic sphenoid sinus encephalocele in an adult by Hunt and Vaicys,⁸ meningocele is most commonly found in the proximal region (occipital meningocele) or may occur in distal part (lumbosacral meningocele). Imaging modalities (computed tomography [CT] and magnetic resonance imaging [MRI]) play a key role for its identification since they aid in prognosis and surgical planning.

To our knowledge, we describe the first case of spontaneous resorption of an occipital meningocele in a full-term newborn boy; the resorption occurred in a few hours. CT had been performed before resorption while MRI was performed after.

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Case Report

A full-term newborn was noted at discharge to have in the occipital region a tubular (~1 cm in length), not skin covered, semitransparent cystic lump, located in the left side (→Fig. 1A). Pregnancy was uneventful with no history of drug abuse, hypertension, diabetes mellitus, stroke, or central nervous system disorders. During pregnancy regular prenatal care was reported and the baby had a spontaneous cry at birth (Apgar score at 5 minutes = 10).

For the presence of this occipital lump the baby underwent CT examination without contrast medium administration. The examination was performed using a specific newborn protocol to reduce the radiation dose. CT showed an enlargement of the cerebellomedullary cistern and temporal and cerebellar liquoral spaces and a very thin defect into the left side of occipital bone that connected the dural space with the skin and the visible lump (→Fig. 1B, C). Moreover, it was noticed a subcutaneous hyperdensity at the level of the visible cystic lump (→Fig. 1D).

Hence, CT demonstrated that the cystic lump contained cerebrospinal fluid and that meninges were herniated outside occipital bone defect and a diagnosis of meningocele was proposed, though skin uncovered meningocele is uncommon. CT proved also the absence of cerebral tissue, thus excluding the diagnosis of encephalocele.

After a few hours, the cystic lump showed complete spontaneous resorption (→Fig. 2).

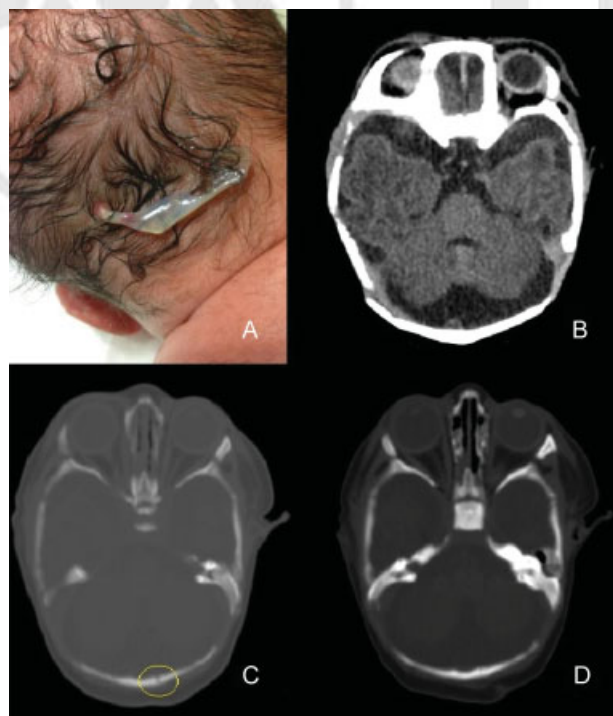


Fig. 1 (A) Semitransparent cystic lump (~1 cm in length), not skin covered, in correspondence of occipital region; (B–D) CT scan shows enlargement of cerebellomedullary cistern and of temporal and cerebellar liquoral spaces (B), the defect into the left side of occipital bone that connected the dural space with the skin (yellow circle) (C) and a subcutaneous hyperdensity in correspondence of visible cystic lump (D). CT, computed tomography.



Fig. 2 Occipital region of the head after complete resorption of the cystic lump.

Anyway, the baby underwent surgery for excision of the lump, to avoid the risk of infection due to the possible communication of the external lump the meningeal space. Postoperatively, he had an uneventful recovery and he was discharged home.

The histology examination was consistent in collagen tissue with meningothelial cells, confirming the diagnosis of meningocele.

After a few weeks, the patient underwent an MRI examination with spin echo T1-weighted, T2-weighted, diffusion-weighted imaging, and gradient recalled echo T2* sequences that confirmed the enlargement of cerebellomedullary cistern, temporal, and cerebellar liquoral spaces (→Fig. 3). No other disorders were found.

Discussion

Meningocele is a very rare disorder and to our knowledge, no case of spontaneous resorption is being reported in the occipital region. It is the rarest form of neural tube defect and occurs at a rate of 1 per 5,000 live births worldwide; it is present both along the spinal cord and skull and in approximately 75% of cases they are occipital in location.^{9,10} Meningocele develops due to failure of closure of the neural tube and is usually covered by pia mater and the arachnoid mater. The dura mater extends up to the margin of the defect and the overlying skin is intact. However, in our case the cystic lump was unusually not skin covered; even though, our imaging diagnosis was histologically confirmed.

Meningocele may be associated with other disorders^{1–3} and, sometimes, it may cause complications, such as recurrent meningitis. An occult involvement of the leptomeninges

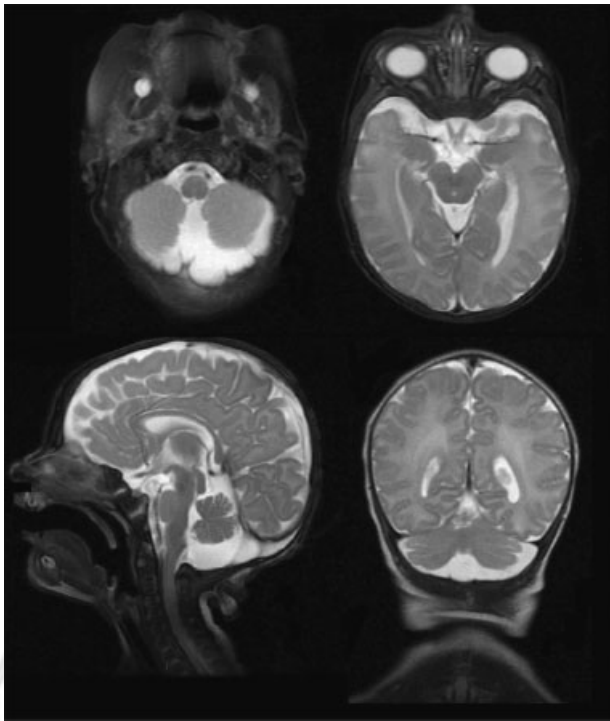


Fig. 3 MRI: axial, coronal, and sagittal T2-weighted sequence with fat-saturation confirmed the enlargement of cerebellomedullary cistern and of temporal and cerebellar liquoral spaces. MRI, magnetic resonance imaging.

may lead to an infiltration by pathological organisms and necessitates restoration of the integrity of the cerebrospinal fluid space to prevent infection. Moreover, meningocele may be complicated by ulceration or rupture.

For these reasons, knowing and recognizing this particular condition is necessary.

Surgery is the preferred treatment used for encephaloceles, and MRI is the best imaging technique to exclude hydrocephalus and to define their content and the extent of cerebral tissue in an encephalocele before surgery.⁹ CT displays the bone anatomy, but the intracranial connection is best defined with MRI.⁹

CT shows the defect into the bone and it allows identifying associated disorders. It also allows having information about the content of the cystic lump.

When encountering a cystic lump in the occipital region a wide spectrum of congenital lesions (e.g., encephaloceles, dermoid and epidermoid cysts, benign tumors) and acquired lesions (e.g., sarcoma, Langerhans cell histiocytosis, metastatic neuroblastoma, infectious or traumatic lesions) have to be considered.⁹ However, in our case, histology proved the presence of collagen tissue with just meningotheial cells, thus confirming the clinical and radiological diagnosis. Immunohistochemistry was not performed since the pathologist was really confident on the diagnosis.

CT scans of the brain can provide information about brain tissue and brain structures, such as the brainstem and ventricular system, and about bone structure, allowing an evaluation of the cranial defect.

MRI is the most accurate imaging examination of the brain: it gives detailed information about tissue and structure; it provides exhaustive indications about the content of the cystic lump in case of meningocele: it is hyperintense on T2-weighted sequences without cerebral tissue in the context. Furthermore, we performed this technique to evaluate eventual associated cranial malformations, since meningocele can be associated with Dandy–Walker syndrome, as previously described in approximately 40 cases in the literature, with Arnold–Chiari malformation, amniotic rupture, cerebellar dermoid tumor, or other disorders.^{3–5}

In our case, MRI was performed after spontaneous resorption and surgical treatment and the cystic lump was not visible anymore. As previously reported by Nejat and Kazmi,⁷ we may presume that this process of reduction may be due to a gradual obliteration of the dural defect and/or a lower pressure after discharge in the meningeal space but we cannot be sure of the two theories.

Treatment for meningocele consists of surgical closure of the protruding sac and a definitive closure of the defect. Considering that meningoceles are frequently covered by a thick epithelial membrane, they often do not present the same surgical urgency as for myelomeningocele.¹¹ In the preoperative preparation, if the lesion is a skin covered antibiotics are not necessary before surgery; if the lesion is open, prophylactic antibiotics should be started in the intensive care nursery.¹¹ Usually, if a meningocele is covered with normal skin, surgery is not urgent and it is recommended to operate when convenient; on the other hand, when it is not skin covered, surgery is considered urgent, because the meningocele can be easily damaged, and may become infected. In our case, considering the gradual perinatal size reduction of the meningocele, the choice of submitting the patient to surgery was debated a lot. However, considering the absence of normal skin covering the lesion, surgical treatment was performed in this newborn to avoid complications, to decrease the risk of infection, and protect the integrity of the brain, due to the cranial skull gap. However, the exact timing of the surgery should be individualized, but the repair performed as early as possible.¹¹

In conclusion, it is important knowing and recognizing this particular condition because it is associated with serious complications; thus, a timely diagnosis is necessary to treat it (also surgically), to avoid the risk of infection, and to recognize any associated disorders. Imaging examinations, in particular CT and MRI, are fundamental for a correct diagnosis.

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